Acute Posterior Multifocal Placoid Pigment Epitheliopathy and “Ampiginous Choroiditis”: Should These Be Treated, And If So, With What?

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What is APMPPE?

- Uncommon condition
- Healthy young adults
  - 30% flu-like illness
- Bilateral, but may be asymmetric
- Rapid onset of blurred VA, photopsias, scotomas (paracentral, central)
- Visual Prognosis – relatively benign, ≤ 20/40 in 58% eyes
Etiology - Unknown

• Associated systemic conditions
  - viral prodrome, Lyme disease, pulmonary TB, gp A strep infection, hepatitis B vaccine, mumps, sarcoid, Wegner’s granulomatosis with polyangiitis (PGA = WG), ulcerative colitis, systemic necrotizing vasculitis

• Associated ocular conditions
  – CRVO, papillitis, periphlebitis, exudative NSD

• HLA association – B7, DR2
Characteristic Fundus Findings

- Multiple flat, yellowish white, placoid lesions, varying in size, at level of RPE
- Lesions fade over time
  - Gradually, with atrophy and hypertrophy of RPE
- New lesions may be observed over several weeks
Evolution of APMPPE Lesions
FA Pattern

Block Early

Stain late
ICG-A  Mendrinos E, Baglivo Eye 2010;24:180 APMPPE – Flu vaccine

- Indocyanine green angiography (late phase) showing irregularly sized hypofluorescent dots unevenly distributed in the fundus in the acute stage of disease (top) that have nearly completely resolved on the follow-up angiogram four weeks later (bottom)
• Fundus autofluorescence (FAF) shows hypofluorescence of the posterior pole placoid lesions
Phases – Hyperacute (1a), Acute (1b)(2-4 d), Subacute (2)(2wks), Late (3)(1.5-6 mo) Resolution (4) (~3 mo)
What is Ampiginous Choroiditis?

Heterogeneity in 6 Cases
What is Ampiginous Choroiditis?

  – 26 eyes of 16 patients
• Yellowish white placoid lesions with geographic borders occurring in the midperiphery and periphery, unlike serpiginous choroiditis. The posterior pole may be involved later in the disease and rarely this may be the initial presentation.
• Lesions are much smaller than serpiginous choroiditis and APMPPE, approximately ½ disc area.
• Lesions are recurrent, unlike APMPPE.
• FA characteristics are markedly different. The active lesions show central hypofluorescence with hyperfluorescent margins. They do not show blocked fluorescence as in APMPPE.
Relentless Placoid Chorioretinitis
TB associated chorioretinitis
Serpiginous Choroiditis – Classic Appearance
After 4.5 Years on Oral Corticosteroids and Azathioprine – VA = 6/9 OU
What is Ampiginous Choroiditis?

• Distinguishing features
  – Distribution of lesions: periphery (mid and far) and macula
  – Morphology of lesions
  – Mild vitritis (35%)
  – Prolong relapsing course: 6 months to 5 years
  – Different complications
    • Present: Subretinal fibrosis and ERM
    • Absent: optic disc swelling, CNV, SRF
Should either of these diseases be treated?

- Occum’s Razor – among competing hypotheses, the one that makes the fewest assumptions should be selected.
- Kaplan’s modification of Occum’s Razor – “treat the cause AND if you don’t know the cause invoke autoimmunity and inflammation as the cause”
APMPPE

• Visual prognosis is generally good and recurrences do not occur
  – Do not treat if VA is $\geq 20/60$
  – If VA is $\leq 20/80$ treat with systemic corticosteroids
    • Bilateral disease
    • May want to withdraw treatment if an infection is masquerading as APMPPE

• Recurrences rarely occur; if they do, think of relentless placoid chorioretinitis and/or ampiginous choroidopathy
Ampiginous Choroidopathy

- View the disease as relentless placoid chorioretinopathy = forme fruste of serpiginous choroidopathy
  - R/O TB
- If center of macula threatened or VA ≤ 20/25 – systemic corticosteroids and IMT
  - Prednisone (1 mg/kg) in four divided doses
  - Azathioprine (1.5 mg/kg) daily
    - Methotrexate or Mycophenolate mofetil
Relentless Placoid Chorioretinitis

Color photographs

FAF